EXACERBATION OF SECONDARY TRIGEMINAL NEURALGIA DUE TO OSTEOMYELITIS IN A CASE OF ROMBERG FACIAL HEMIATROPHY

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ABSTRACT

Progressive facial hemiatrophy or Romberg syndrome, is a sporadic disease of unknown etiology characterized by progressive and self-limited shrinking and deformation of one hemiface accompanied by various neurologic complication, including trigeminal neuralgia. We present the case of a 31-year-old caucasian female with exacerbation of a left trigeminal neuralgia two weeks after a dental extraction. The severe pain was due to ostheitis of the left mandible and a fracture through the 38 alveola. After removing the necrotic tissue and mandibulo-maxilary fixation the evolution was favorable with no signs of infection or wound dehiscence and the symptoms of pain were relieved.

Key words: progressive facial hemiatrophy, Romberg syndrome, trigeminal neuralgia, osteomyelitis

BACKGROUND

Progressive facial hemiatrophy (PFH), or Parry-Romberg or Romberg syndrome, is a sporadic disease of unknown etiology characterized by progressive and self-limited shrinking and deformation of one hemiface which involves different tissues, scar-like cutaneous changes with subcutaneous connective and fatty tissue atrophy, circumscribed osteoporosis, bone deformation with hyperpigmentation of the overlying skin and it could be accompanied by various neurologic complication, including migraine-type headache, trigeminal neuralgia, and focal refractory epilepsy.

The disease most commonly occurs during the first or second decade of life, when facial and head growth might be most affected, with a higher incidence rate in females.

Patients who manifest atrophy in early ages, have a better outcome.

The disease is likely to have an acquired autoimmune mechanism. Progressive facial hemiatrophy may be a form of focal scleroderma. The diagnostic differentiating feature between the two is the preservation of elastic tissue at histologic examination in progressive facial hemiatrophy. Encephalitis, complicated migraine, and slow virus infection have also been advanced as possible causes.

Several authors have proposed that progressive facial hemiatrophy results from a disturbance leading to sympathetic nervous system hyperactivity, the trophism of the fat and subcutaneous tissues being under the influence of the sympathetic nervous system.

Recently blink reflex and trigeminal evoked potential studies have indicated abnormalities in brain stem. Hyperactivity of the brain stem sympathetic centers, possibly caused by an autoimmune process may be the primary cause for cutaneous and subcutaneous atrophy in Parry-Romberg Syndrome. This disease may be due to neural crest defect.

The extension of the atrophy is frequently limited to one side of the face, and the ipsilateral involvement of body is rare. Ocular involvement is

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common in the form of enophthalmous, pupillary disturbance, heterochromia, uveitis, pigmentary disturbance of ocular fundi. There may be associated otorhinolaryngological disorders in the form of unilateral hearing loss, frontal sinus deformity and parotid gland atrophy, dental abnormalities and trismus/jaw symptoms (hemimasticatory spasm). Parry Romberg disease may be associated with hemiatrophy of brain, breast and whole half of the body along with pigmentary skin abnormalities such as vitiligo, facial nevus and morphoea along with sweating and lacrimation abnormalities.

Migraine and facial pain are the commonest neurological symptoms in this patient group. Occasionally, epilepsy may occur and can sometimes be related directly to a brain abnormality ipsilateral to the skin lesion. There are a range of other cerebral abnormalities such as cerebral hemiatrophy, meningeal thickening, cortical dysgenesis, calcified lesions, aneurysms, and intracranial vascular malformations. There is one reported fatal case in a child with associated "Rasmussen-type" encephalitis.

CASE STUDY

We present the case of a 31-year-old caucasian female who was admitted to the Oral and Maxillofacial Surgery Department with a mild swelling of the left mandibular region, hyperemia, and pain when she attempted normal functional movements of the jaws.

The patient was previously hospitalized in the neurology clinic for exacerbation of a left trigeminal neuralgia, especially in the mandibular division, in the past two weeks after a dental extraction. The patient had medical history of left Romberg hemifacial atrophy beginning at age 22 and for about two years the patient had been suffering from episodes of neuralgic pain on the left side of her face lasting from two to fourty minutes, mainly in the temporal and mandibular zone. The patient had no other medical history and family records were noncontributory, with no reports of trauma, use of medication or major illness.

At the admission to the neurology clinic on physical examination, there was unilateral linear atrophic, fibrotic, and hyperpigmented plaques with remarkable wasting of underlying soft tissue, muscle and bone on the left side of her face and prominence of the zygomatic arch, and of all performed measurements only the distance between mandibular angle and menton was 9 cm on the left side and 8 cm on the right side. The neurologic exam was normal.

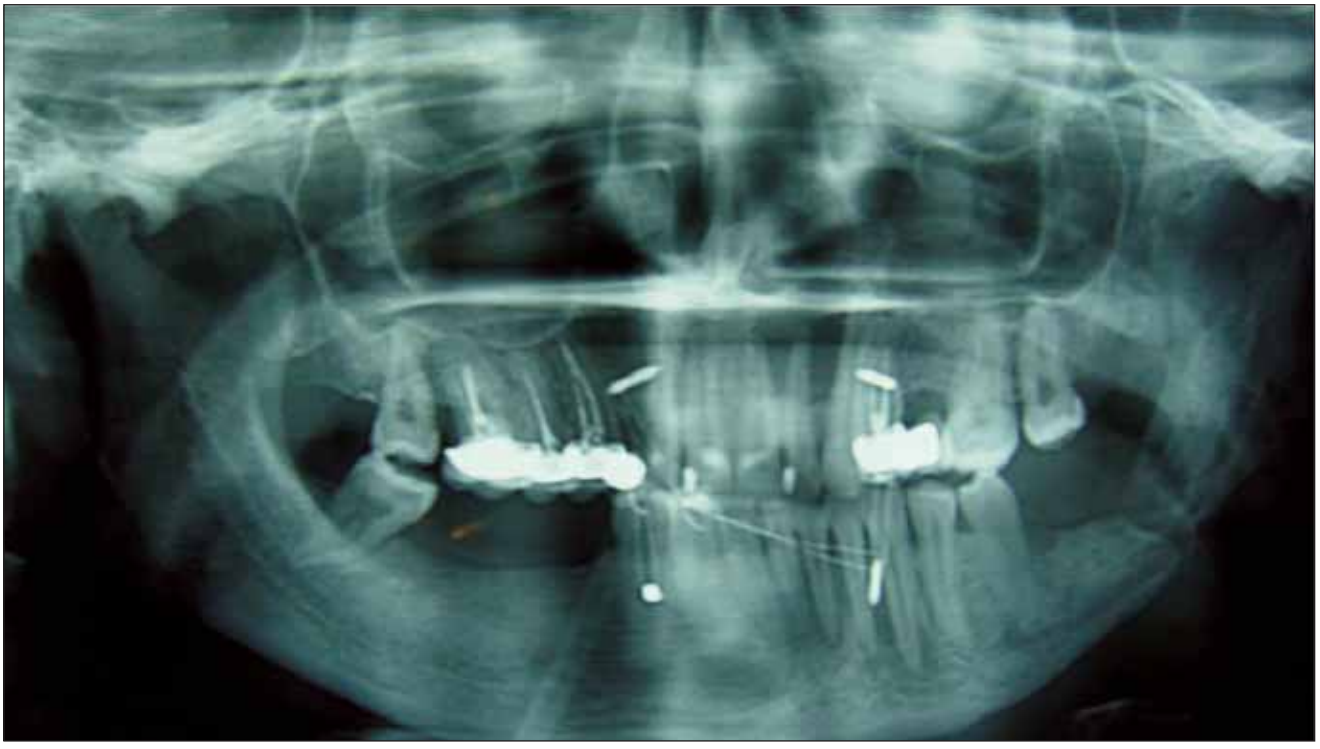
The laboratory blood tests (hematologic screening, routine biochemical profile, urine analysis) were normal except for a mild leucocytosis with neutrophilia, and so did the chest roentgenography and 12-lead electrocardiography.

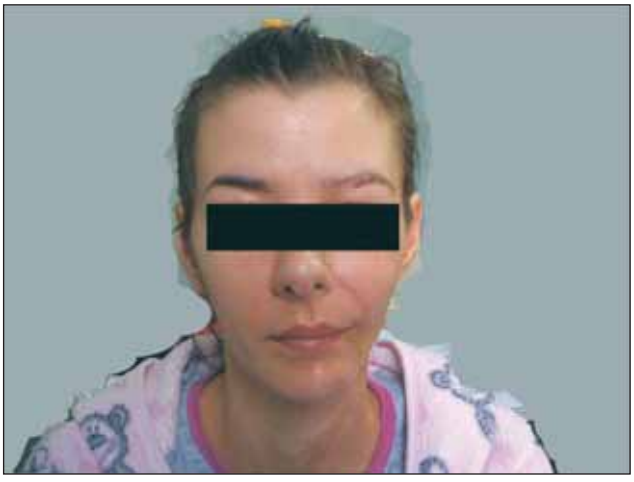
The cerebral CT scan and electroencephalography were also normal.

The oral exam revealed a diminished opening of the mouth, alveolitis at the site of the extracted 38 and periosthitis of the left mandibular corpus. The

panoramic x-ray revealed ostheitis of the left mandible and a fracture through the 38 alveola. Under general anesthesia the necrotic tissue from the site of the fracture was removed, a local flap was designed in order to cover the open alveola, but instead of rigid fixation was practiced mandibulomaxilary fixation. We adopted this method in order to minimize the risk of infection at the site of the fracture. There were administrated antibiotics and non steroid antiinflamatories for seven days.

The evolution was favorable there was no sign of infection or wound dehiscence, the symptoms of pain were relieved. After four weeks the intermaxilary fixation was removed, and there was no sign of complications at the site of the fracture. The panoramic x-ray performed three months later showed that the two fragments are continuous, there is no displacement, and the occlusion is as before the fracture.





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